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Case Study

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ROSS SYNDROME – CASE SERIES

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ABSTRACT

Ross syndrome is rare neurocutaneous disorder which characterised by a triad od segmental anhidrosis, areflexia and tonic pupil. This syndrome is not easily diagnosed and required clino-pathological correlation. Herby we present a case series of Ross syndrome during one year of study period.

KEYWORDS: Anhidrosis, Areflexia, Harlequin syndrome.

INTRODUCTION

Ross syndrome, named after AT Ross who first described it in 1958, is a rare neurocutaneous disorder characterized by a triad of segmental anhidrosis, areflexia, and tonic pupils.[1] Harlequin syndrome encompasses the first component of the triad, while the latter two components define Holmes–Adie syndrome, presenting a pathogenetic relationship that complicates the differentiation from Ross syndrome.[2] This syndrome is often challenging to diagnose due to its low prevalence and variable clinical manifestations, with less than 100 reported cases in literature. The disorder's unpredictable course adds to the diagnostic challenge, and its causation has been attributed to various factors such as autonomic denervation, autoimmunity, developmental origin, viral infections, and genetic factors.[3][4][5] The underlying pathology involves dysfunction of the sympathetic and parasympathetic nervous systems, leading to a range of autonomic dysregulations.[6]

CASE SERIES

The following cases were received at Department of Pathology SMS Medical College Jaipur. Haematoxylin and Eosin staining were performed on the biopsy sample received.

Case 1

Patient history: A 24-year-old male presented with insidious onset of anhidrosis on the right half of the body. Skin biopsy samples were received for analysis from both sides of the body. Morphological and quantitative assessment of sweat glands was done.

Case 2

A 35-year-old male patient presented with complaints of excessive sweating on right trunk and right lower limb lasting for 5 years associated with anhidrosis over left side chest and left upper limb. Skin biopsy samples from both sides of the body were received.

Case 3

A middle aged homemaker presented with absence of sweating on the right side of the body for the last 7 years with increasing tiredness and heat intolerance for 2 years. She also had excessive sweating over left side. Skin biopsy samples from sides of the body were received.

DISCUSSION

Panda et al. conducted a study revealing distinctive histopathological findings associated with Ross syndrome. Their investigation demonstrated focal flattening of rete ridges in the epidermis, coupled with a relative increase in eccrine glands at the dermis and subcutaneous junctions in regions characterized by hyperhidrosis. Conversely, areas exhibiting hypohidrosis displayed a relative decrease in eccrine glands at the lower dermis.[7] Similar findings were noted by Metta AK, where histopathological examination of the anhidrotic area in Ross syndrome revealed sparse to absent eccrine sweat glands, providing histological evidence for the impaired sweat response in these regions. Conversely, biopsied skin from the hyperhidrotic area exhibited an increased number and size of eccrine sweat glands, accompanied by epidermal hyper melanosis.[8] These histopathological observations provide valuable insights into the skin alterations associated with the dysregulation of sweating in Ross syndrome, contributing to a deeper understanding of the pathological mechanisms underlying this rare neurocutaneous disorder. The hypohidrosis observed in Ross syndrome is attributed to the damage inflicted upon postganglionic sympathetic fibers that innervate sweat

glands. This damage results in a reduced network of fibers lacking receptors for vasoactive intestinal peptide (cholinergic) or dopamine-β-hydroxylase (noradrenergic) axons. In contrast, the phenomenon of excessive sweating, a distressing symptom exacerbated by exercise and elevated temperatures, is considered compensatory or a consequence of the loss of cholinergic M2 inhibitor presynaptic auto receptors. Notably, the persistence of excessive sweating may eventually evolve into anhidrosis, further emphasizing the dynamic and evolving nature of the autonomic dysregulation seen in Ross syndrome.

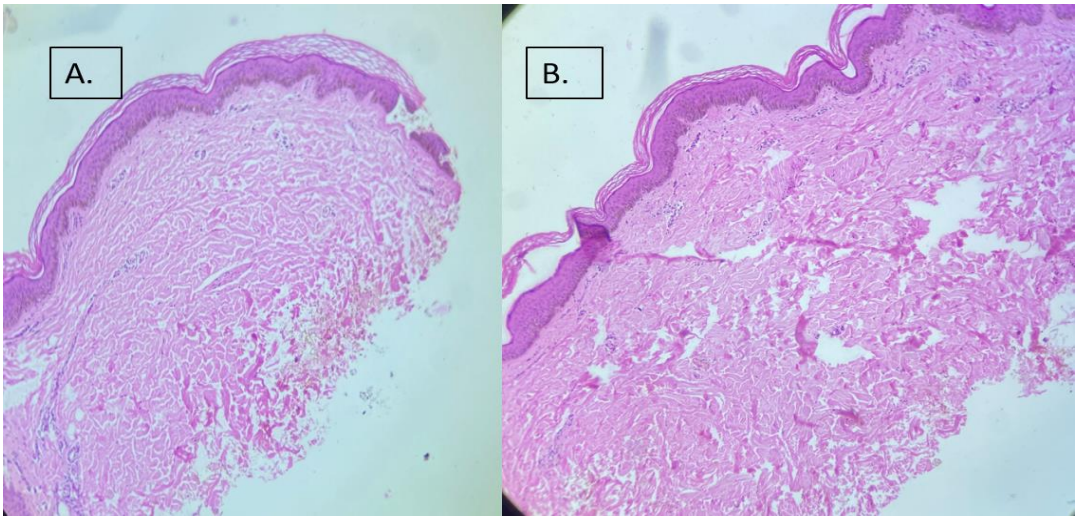

Figure 1: 10x HP image showing skin punch biopsy with hypohidrotic region (without eccrine gland).

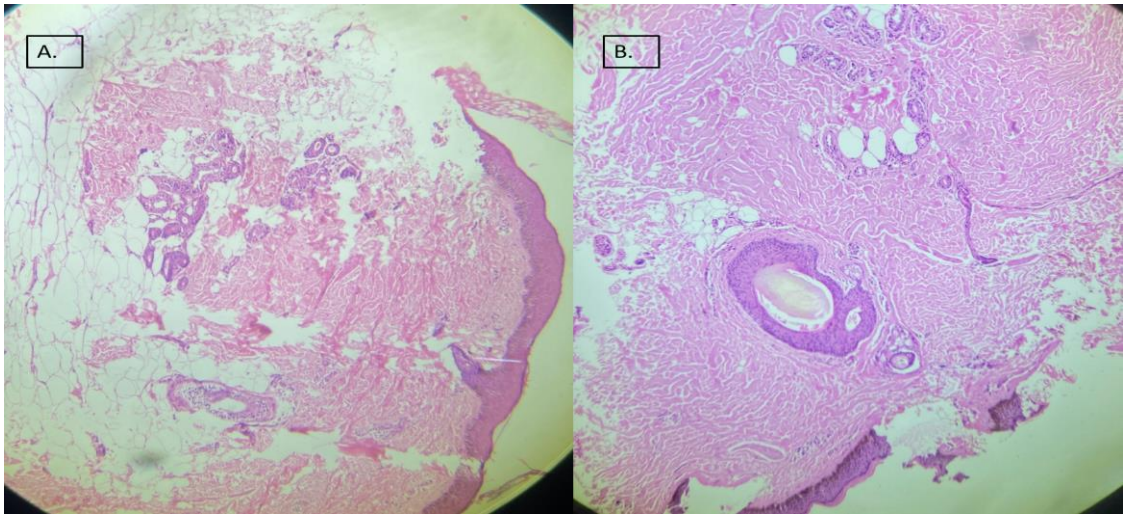

Figure 2: 10x HP image showing skin biopsy with hyperhidrotic area.

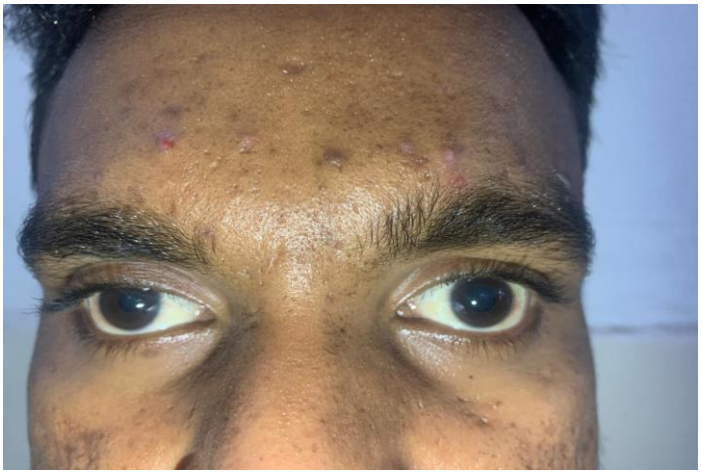

Figure 3: Sluggish reacting pupil on left side with dilation lags which reacted better to accommodation.

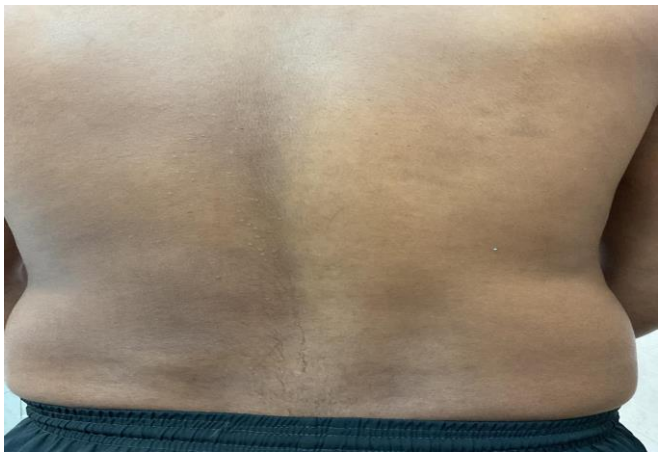

Figure 4: Anhidrosis over left side of back.

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None.

Conflict of interest
None.

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